Structural Vascular Pathology in Children

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Intracranial Aneurysms in the Pediatric Population

Developmental Features and Pathology

- Congenital and acquired influences.
- Degenerative changes at the arterial bifurcation caused by axial blood flow.
- Similar wall features to adult aneurysms: Absence of the internal elastic lamina and the muscularis layer of the media.

Developmental Features and Pathology

- Infection and trauma are more common in pediatric aneurysms.
- Associated disorders: Coarctation of the aorta, polycystic kidney disease, tuberous sclerosis, and HIV infection.
- Strong predilection for the ICA bifurcation.
- Posterior circulation aneurysms are three times more prevalent in children.
- Giant aneurysms are more common.

Epidemiology

- Incidence: 1-3 cases / 1 million pop. / year
- Male / Female : 1.8 / 1
- Cooperative Study (1966): 41 of 6368 ruptured aneurysms (0.6%).
- More recent studies: 1 % of all surgically treated aneurysms. Proust et al. J Neurosurg 2001;94:733-9

- Most academic centres treat one / year.

Intracranial aneurysms in the pediatric population: case series and literature review

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- 706 cases have been described since 1939.
- Male/female preponderance of 1.8:1
- SAH is the most common presentation: ruptured aneurysms are 4 times more common than unruptured aneurysms.
- Anatomic location at the ICA bifurcation is a distinguishing feature of aneurysms in this population: 26 % v/s 4.5% in adults (Cooperative Study).
- Posterior fossa aneurysms: 17% of all intracranial aneurysms. (Adults: 8% Cooperative Study).
- Giant aneurysms: 20% of all intracranial aneurysms.
- Infectious aneurysms: 2%

Huang et al. Surgical Neurology 2005:63:424-433

Clinical Assessment

- The management strategies follow the same rules than in adults.
- Most common presentation (52%-74%):
  Sudden headache.
  Vomiting.
  Seizures.
  Decreased LOC.
  Meningismus.
- Rebleeding is common: 57% of cases; half occurs within the first 24 hours.
- Children are less vulnerable to vasospasm.

Investigations

- Infections should always be in the differential diagnosis : LP.
- CT
- CTA
- MRI
- MRA
- DSA.

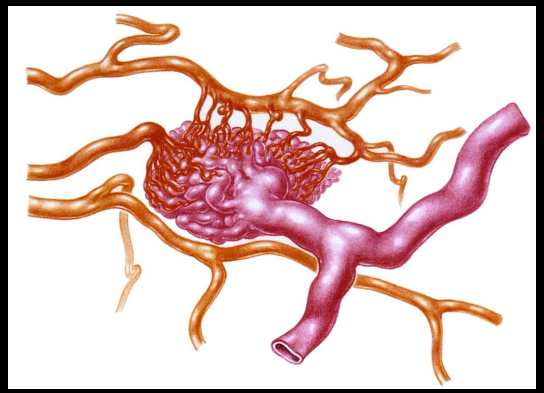
Definitive Treatment

- Surgical clipping: Delayed fashion?
- Endovascular treatment: coiling, balloon occlusion.

Pediatric Arteriovenous Malformations

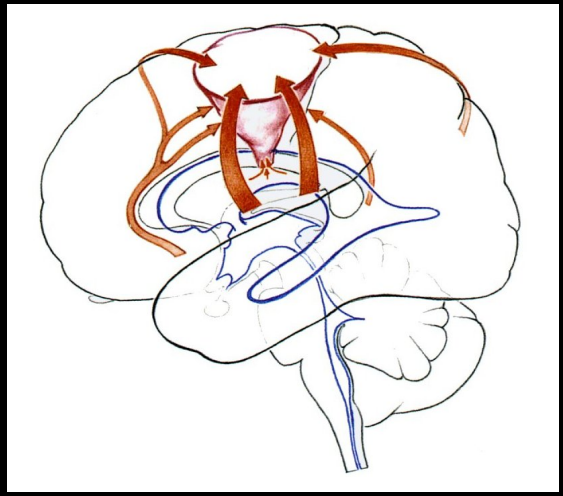
- Congenital vascular malformations.
- High flow lesions that consist of a mass of arteries and arterialized veins fed from the carotid or vertebrobasilar system.
- Histology: arteriovenous shunts consisting of

1) Feeding arteries.
2) Mass of coiled vessels (the nidus).
3) Venous sinusoid spaces and veins communicating directly without an interposed capillary network.



Developmental Features and Pathology

- Structural defect in the formation of the primitive arteriolar-capillary network.
- Most occur before 40 mm embryonal length.
- Expansion occurs by enlarging and increasing the tortuosity of the feeding and draining channels.
- Course to the ventricle in an inverted wedge-shaped fashion.
- Potential role of postnatal factors.



Epidemiology

- Incidence: 1 per 100,000 children
- 12% - 18% become symptomatic during childhood.

- Location: Posterior fossa ⟶ 10%.
  Midline ⟶ 5%-10%.
  Supratentorial and lateralized ⟶ 80%-85%.
- Multiple AVMs: 17%

Clinical Presentation

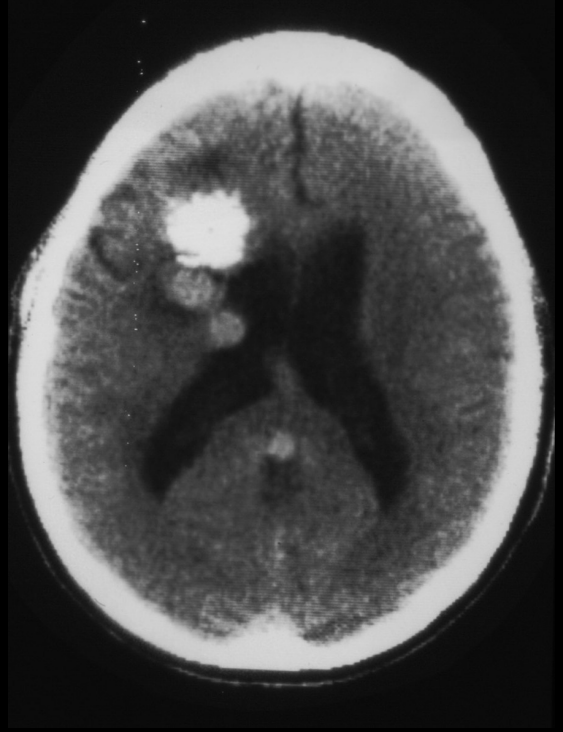
- Hemorrhage: Most common presentation (50%-80%).
- Seizures (12%-25%).
- Congestive heart failure (18%).
- Headache
- Progressive neurological manifestations and mental deterioration – arterial steal phenomenon

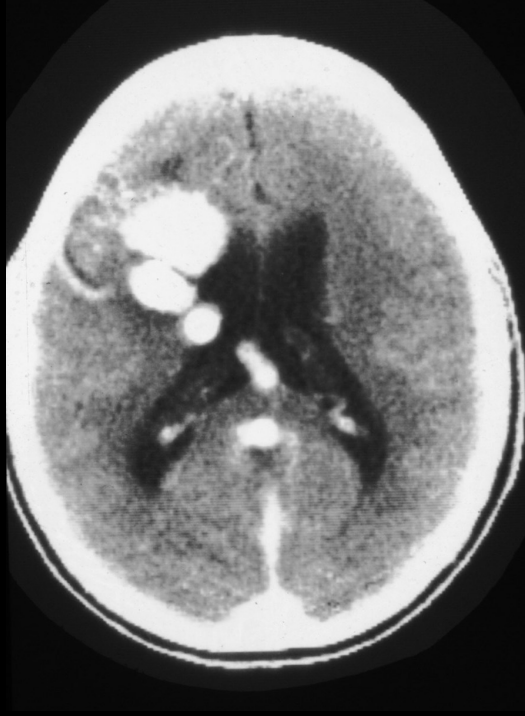
Natural History

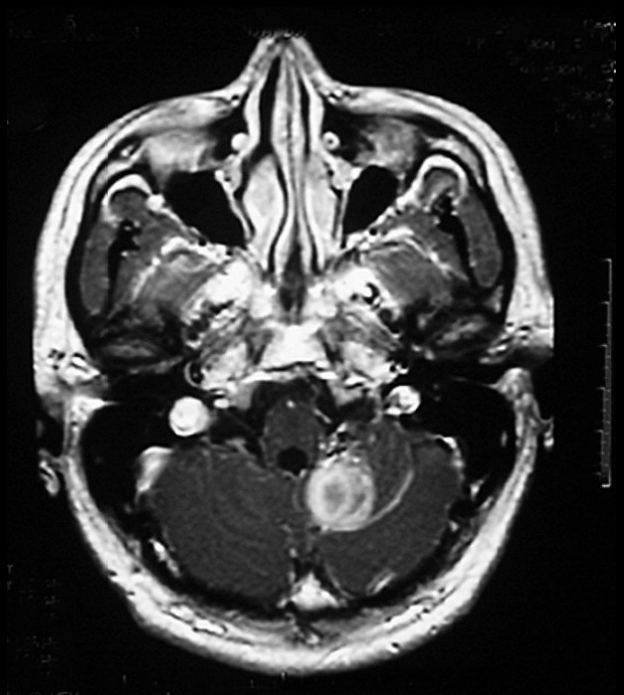
- It is not well known.
- Some evidence of higher risk of bleeding and rebleeding.
- Rebleeding rate at 5 years:
  Adults → 13%.
  Children → 25%.
- Possible explanations:
  Higher number of post. fossa AVMs.
  "Younger vessels" more fragility.
  Children's AVMs tend to be smaller.
- Mortality:
  First hemorrhage: 24%.
  Second hemorrhage: 13%-20%.
- Recurrence.

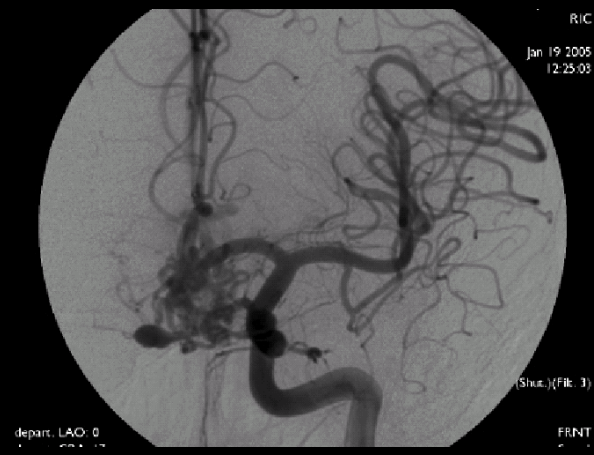
Investigations

CT
CTA
MRI
MRA
DSA
U/S Neonates









Treatment

- Considering the high mortality associated with the natural history, the aim of therapy is the complete removal.
- Surgery:
  Acute period: Evacuation of hematoma. Treatment of increased ICP – EVD.
  Delayed surgical resection: 3-7 days after the hemorrhage. Rebleeding v/s Vasospasm.

Results of Microsurgery for Cerebral AVMs in Childhood

Complete resection: 60% of the cases

Outcome: Excellent or good result in more than 85% of the children

Mortality rate: 3%

Menovsky et al. Eur J Pediatr 1997;156:741-746

Author	No. of Cases	Age (years) (range)	Location	Outcome	Mortality
Long et al. [33]	8	0–5	Supratentorial AVMs	No neurological deficit	25% (due to cardiac arrest)
Kelly et al. [24]	2	<1	Supratentorial AVMs	Very good, no neurological deficit	0%
	17	1–20	Supratentorial and infratentorial AVMs	Very good ($n = 12$), minor deficit ($n = 4$, already preexsistent)	6%
Amacher et al. [2]	20	<18	Supratentorial and infratentorial AVMs	75% no neurological deficit	0%
Mori et al. [38]	18	0–15	Supratentorial and infratentorial AVMs	Normal ($n = 4$), slight neurological deficit ($n = 3$), moderate neurological deficit ($n = 2$), poor ($n = 3$)	22%
Laine et al. [28]	20	<20	Supratentorial and infratentorial AVMs	60% normal, 20% disabled	20%
Gerosa et al. [12]	38	2–16	Supratentorial and infratentorial AVMs	Good ($n = 22$), fair ($n = 9$), total excision in 23 patients	8%
Mazza et al. [35]	18	<16	Supratentorial and infratentorial AVMs	83% normal, 6% disabled	11%
Humphreys et al. [18]	3	10–13	Brainstem AVMs	Very good, neurological deficit in 1 patient	0%
Celli et al. [4]	11	9–15	Supratentorial and infratentorial AVMs	Excellent ($n = 4$), good ($n = 3$), fair ($n = 1$), poor ($n = 2$)	0%
Humphreys [17]	72	< 17	Supratentorial and infratentorial AVMs	60% no neurological deficit, 32% neurological deficit, total excision in 54 patients	8%
Eiras et al. [8]	15	0–16	Supratentorial and infratentorial AVMs	Normal ($n = 9$), abnormal ($n = 2$), total excision in 12 patients	0%
Garza-Mercado et al. [11]	12	9–18	Supratentorial and infratentorial AVMs	Neurological deficit ($n = 5$), incomplete excision in 3 patients	25%
Yasargil [56]	60	< 18	Supratentorial and infratentorial AVMs	90% normal	0%
Fong and Chan [9]	27	0–16	Supratentorial and infratentorial AVMs	85% neurological, normal	4%
Kahl et al. [22]	33	1–16	Supratentorial and infratentorial AVMs	No worsening of neurological signs, complete excision in 31 patients	3%
Suarez and Viano [49]	10	< 15	Supratentorial and infratentorial AVMs	70% neurological, normal	30%
Lapras et al. [29]	62	< 15	Supratentorial and infratentorial AVMs	42% normal, 40% neurological deficit	10%
Malik et al. [31]	27	0–18	Supratentorial and infratentorial AVMs	Excellent ($n = 17$), good ($n = 6$), fair ($n = 2$, conform pre-operative status)	7%
Kondziolka et al. [25]	97	0–18	Supratentorial and infratentorial AVMs	Normal neurological outcome in 70%, seizure control in 73%	5%
U et al. [53]	2	12–14	Deep periventricular AVMs	Good-excellent	0%
Lasjaunias et al. [30]	8	< 15	Supratentorial and infratentorial AVMs	Moderate neurological deficit ($n = 2$)	0%

Treatment

- Intravascular Embolization:
  Cure is seldom obtained with embolization alone (5% of cases).

  Staged embolizations and pre-operative embolization – Large MAVs.
  Reduction of size prior to radiosurgery.

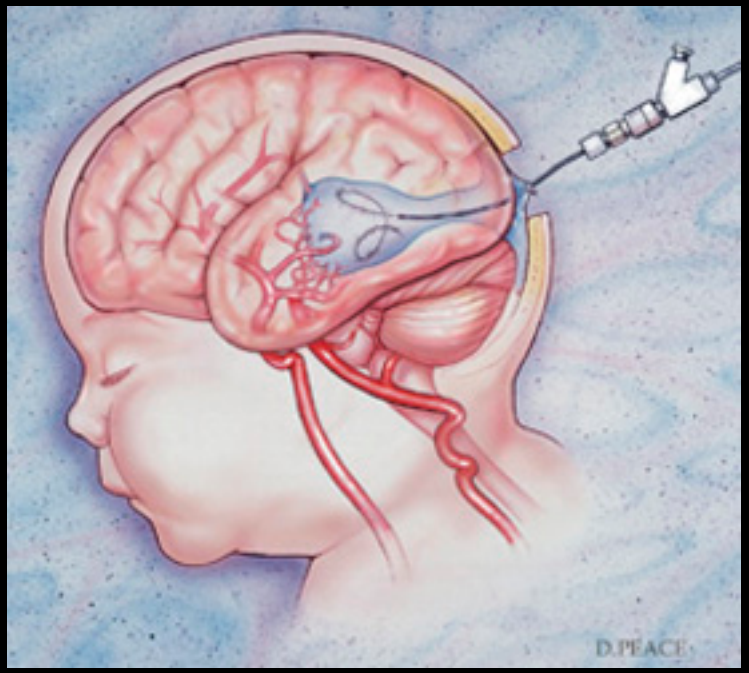
Frizzel et al. Neurosurgery 1995;37(6):1031-1040

Radiosurgery

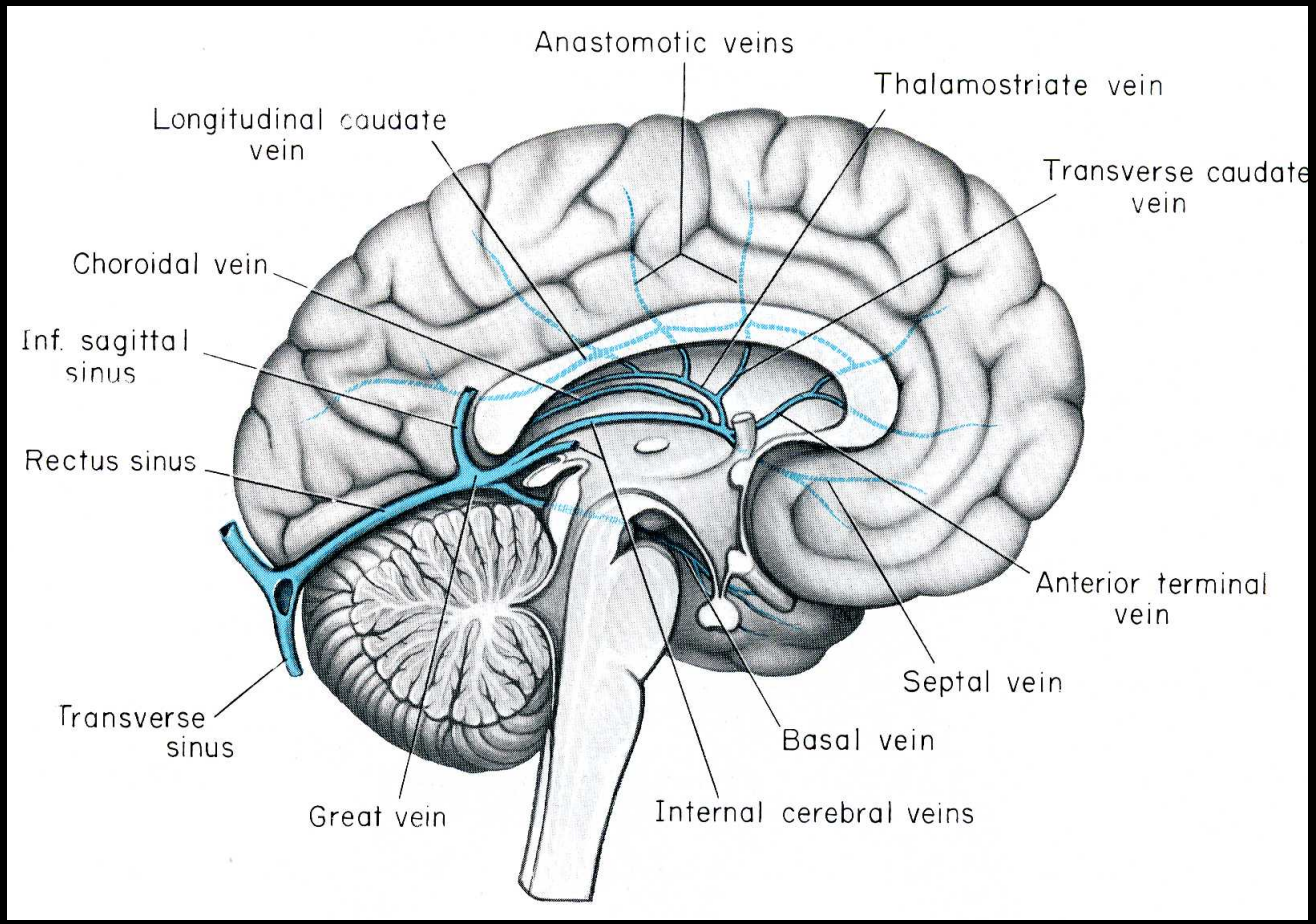
- Safe and effective alternative for small (< 30 mm) and inoperable AVMs.
- Latency: months to years.
- Long-term effects in children are not completely known.

Multimodality treatment

Vein of Galen Malformations



Anatomy



VOG is located in the quadrigeminal cistern under the splenium of the corpus callosum.

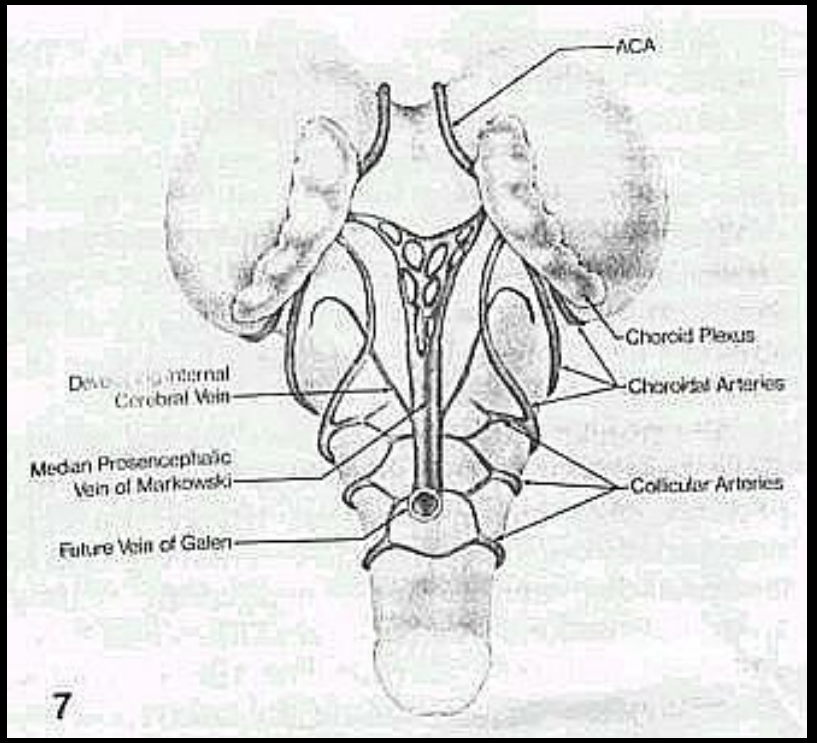
Drains: thalamus, occipital lobes, medial temporal lobes and superior cerebellar vermis.

History

- Galen (second century A.D.) described the deep venous system in lower animals.
- Steinheil (1896) described the first VOG malformation.
- Jaeger, Oscherwitz and Davidoff, and Boldrey and Miller: First clinical and surgical attempts.
- Gold (1964) developed the first clinical classification.
- Hoffman (1982) compiled all the data available in the literature (128 cases). His conclusions:
  Surgical treatment is better than conservative management.
  Infants and older children did better than neonates.
- Lasjaunias (1987) developed a simple radiographic classification.
- Yasargil (1988) proposed a radiographic classification.
- Endovascular therapy (1980's).

Embryology

- Primary VOG malformations: Abnormal remmants of the prosencephalic vein.
- Secondary VOG malformations: Adjacent dural fistulas or parenchymal AVMS draining and dilating a normally developed VOG.



Pathophysiology

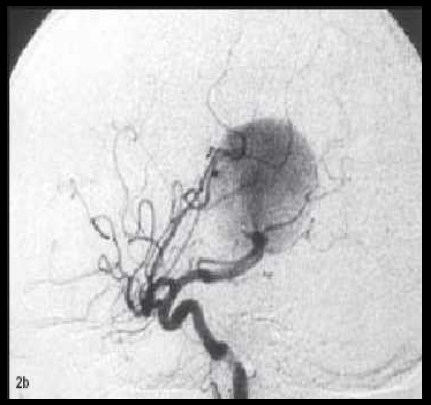
- These AV shunts in utero can result in:
  High-output cardiac failure.
  Cerebral venous hypertension → hydrocephalus, chronic cerebral ischemia and cerebral hemorrhage.
  Pulmonary venous hypertension.

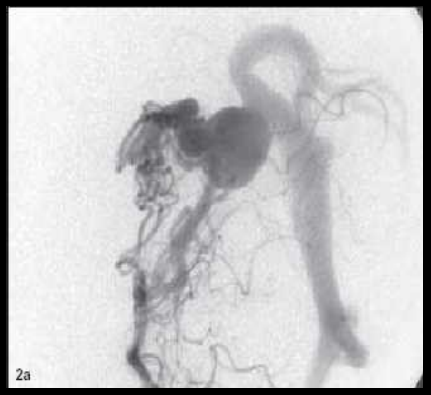
Clinical Presentation

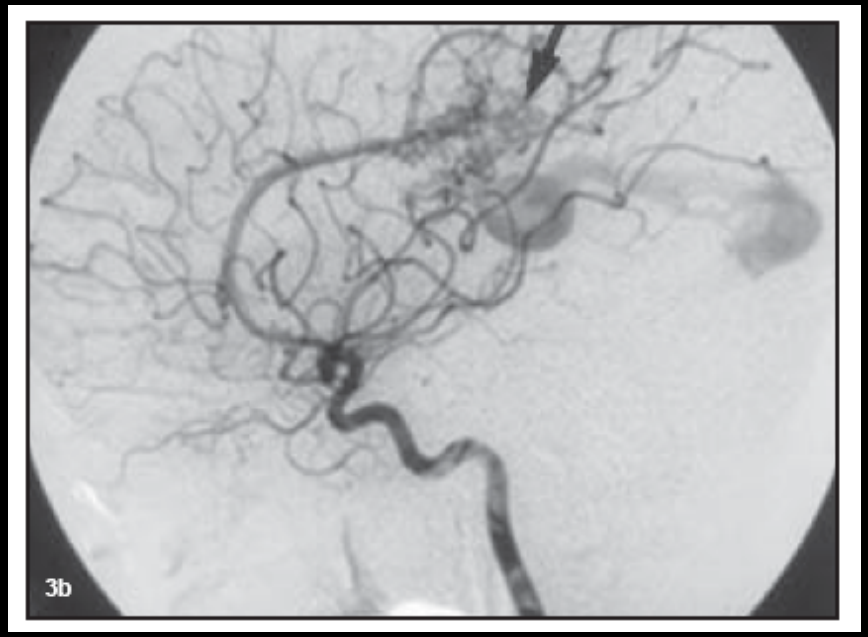
- The clinical presentation and outcome are age related.
- Neonates: Major injury to the brain.
  Medically refractory cardiac failure.
  Urgent reduction of the shunt.
- Infants: Cardiac failure is usually controlled with medical treatment.
  Mild to moderate intracranial venous hypertension with enlargement of the head.
- Older children and young adults: Headache.
  Learning disabilities.
  Increasing spasticity.
  Seizures.
  SAH.

Lasjaunias Classification

- Primary or true VOG malformations:
  Mural.
  Choroidal.
- Secondary







Yasargil Classification

- Type I: Posterior cerebral artery / pericallosal artery. No thalamoperforating arteries.
- Type II: Thalamoperforating arteries.
- Type III: Mixed pattern; pericallosal, thalamoperforating, and posterior cerebral arteries.
- Type IV: Secondary type of Lasjaunias.

Treatment

- Treatment is better than conservative management.
- Combined endovascular and surgical approaches offer the best outcome.
- Endovascular treatment requires transvenous and transarterial embolization in a staged fashion.
- Open surgery is reserved for:
  Patients with hydrocephalus.
  Exposure during transtorcular embolization.
  Malformations not amenable for embolization.

Treatment

- Vein of Galen malformation with limited number of feeders (Type I, Mural)
- - endovascular embolization (arterial approach) is treatment of choice
- - preserves venous drainage
- - multiple stages may be required

- Type II/III, Choroidal
- - arterial approach preferred
- - transfemoral venous endovascular approach

- True AVM (Type IV, Dilatations)
- - multimodality involving endovascular, surgery, radiosurgery